Vulval Disorders; Update

Prof. Amr El-Shalakany, MD FRCOG.
Gynecological Oncology Unit
Early Cancer Detection Unit
Ain Shams University
Cairo, Egypt

Vulval Disorders

- Vulval complaints form 5-10% of all gynaecological complaints
- Tendency for under reporting and less attention from physicians
- Can cause extreme distress and may lead to marital problems

Vulval Disorders

- The vulva is a piece of skin,
- Therefore, general dermatologic conditions (e.g. eczema, psoriasis, pemphigus, lichen simplex, lichen planus) must be considered when faced with a vulval lesion.

The vulva

***It is of paramount importance to involve dermatologists and genito-urinary physicians where the diagnosis is in doubt.

***Always remember that a vulval complaint can be a manifestation of a systemic disease e.g. diabetes, psoriasis, SLE.

Systemic diseases

Vulval diseases

Lichen planus

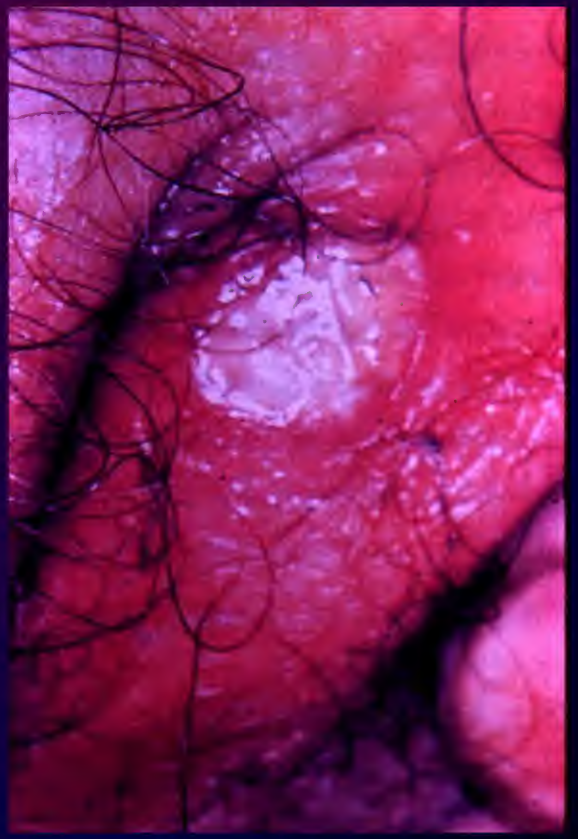
Vulval candidiasis

Childhood vulvovaginitis

Vulval H. simplex & Zoster

Vulval Mollscum contagiosum

Behcet's syndrome



Vulval haemangioma

Vulval lymphoma

Vulval Choriocarcinoma

Vulvar lesions

Fibroepithelial polyp	**Granulomatous Cheilitis** Melkersson-Rosenthal disease

Confusing findings

Indications for colposcopy of the vulva:

1. Changes in skin pigmentation.
2. Ulcers, warts, nodules,thickening, fissuring.
3. Pruritus vulvae, vulval burning, vulvodynia.
4. Dysuria (in the absence of UTI or calculi).
5. Follow up of treated VIN, non neoplastic disorders or vulval carcinoma.

Colposcopy of the vulva is more difficult to interpret than that of the cervix or vagina.

***Keratinization of vulval skin can mask or alter the appearance of underlying epithelium and vasculature.**

***Therefore, identifying lesions and grading their severity is more complex.**

**The junction between the glycogen bearing vaginal epithelium and the keratinized vulval skin is at high risk for developing VIN.

**If VIN or HPV is suspected, it is obligatory to proceed to colposcopic examination of the cervix and vagina, including collection of a cervical smear for cytology.

Technique of vulval colposcopy:

1. Explain the technique to the patient.
2. Modified lithotomy position.
3. Inspection with the naked eye.
4. Paint the vulva with hydrogel.
5. Inspection with low power magnification (6-fold) in a systematic order.
6. Application of 5% acetic acid for 3 min. then repeat inspection.

Toluidine blue application is unreliable for the detection of lesions.

Vulval biopsy:

It is mandatory to confirm the histopatologic nature and the severity of the lesion.

Biopsy techniques are:

- **Excision biopsy (whole lesion is removed)**
- **Keye's punch biopsy.**
- **Large Knife incision biopsy.**
- **Diathermy loop biopsy.**

Vulval biopsy (Keye's punch biopsy)

Classification of vulvae disease (1987):

(1) Non neoplastic vulvae epithelial disorders:

I. lichen sclerosus .

II. Squamous cell hyperplasia.

III. Other dermatosis.

(2) Vulval intraepithelial neoplasia (VIN).

(3) Human papilloma (HPV) virus infection:

I. Condyloma accuminata.

II. Subclinical HPV infection.

ISSVD Classification of Vulvar Dermatoses (2006) :

- **Spongiotic pattern**
  - Atopic dermatitis
  - Contact allergic dermatitis
  - Irritant contact dermatitis
- **Acanthotic pattern** (formerly squamous cell hyperplasia).
  - Psaoriasis
  - Lichen simplex chronicus (primary and secondary)
- **Lichenoid**
  - Lichen sclerosus
  - Lichen planus

ISSVD Classification of Vulvar Dermatoses (2006) :

- **Dermal homogenisation / sclerosis**
  - Lichen sclerosis
- **Vesiculobolbous pattern**
  - Pemphigoid
  - Linear Ig A disease
- **Acantholytic pattern**
  - Hailey-Hailey disease
  - Darier disease
  - Papular genitocrural acantholysis

ISSVD Classification of Vulvar Dermatoses (2006) :

- **Granulomatous**
  - Crhon's disease
  - Melkersson-Rosenthal disease
- **Vasculopathic pattern**
  - Behcet's disease
  - Aphthous ulcer
  - Plasma cell vulvitis

ISSVD terminology and classification of vulvar pain 2003

- **Vulvar pain related to a specific disorder**
  - Infectious (e.g. candidiasis..)
  - Inflammatory (e.g. lichen planus…)
  - Neoplastic (e.g. carcinoma…)
  - Neurologic (e.g. Herpes, nerve compression)
- **Vulvodynia (pain not related to a specific disorder)**
  - **Generalised**
    - Provoked (sexual or non sexual)
    - Unprovoked
    - Mixed
  - **Localised** (vestibulodynia, clitorodynia, hemivulvodynia)
    - Provoked
    - Unprovoked
    - Mixed

Vulvodynia

- Most likely, there is not a single cause.
- Embryologic abnormalities,
- Increased urinary oxalates,
- Genetic or immune factors,
- Hormonal factors,
- Inflammation, infection,
- Neuropathic

Vulvar Care Measures

- Wearing cotton underwear in the daytime and none at night.
- Avoiding vulvar irritants (perfumes, dyed toilet articles, shampoos, detergents, and douches).
- Use of mild soaps, with none applied to the vulva.
- The vulva can be cleaned gently with water and patted dry.
- Emollient without preservatives (vegetable oil or plain petrolatum) helps to hold moisture in the skin and to improve the barrier function.

Vulvar Care Measures

- If menstrual pads are irritating, cotton pads may be helpful.
- Adequate lubrication for intercourse is recommended.
- Ice packs are helpful in some, but produce irritation when overused.
- Cool gel packs may be used.
- Rinsing and patting dry the vulva after urination may be helpful.
- Use of hair dryers should be avoided.

Vulvodynia

- Vulval care measures
- Topical emolients, steroids, local anaesthetics, estrogens
- Oral antidepressants, anticonvulsants, SSRIs
- Low-Oxalate Diet with Calcium Citrate Supplementation
- Botulinum toxin
- Nitroglycerine
- Biofeedback, hypnotherapy, acupuncture
- Vestibulectomy

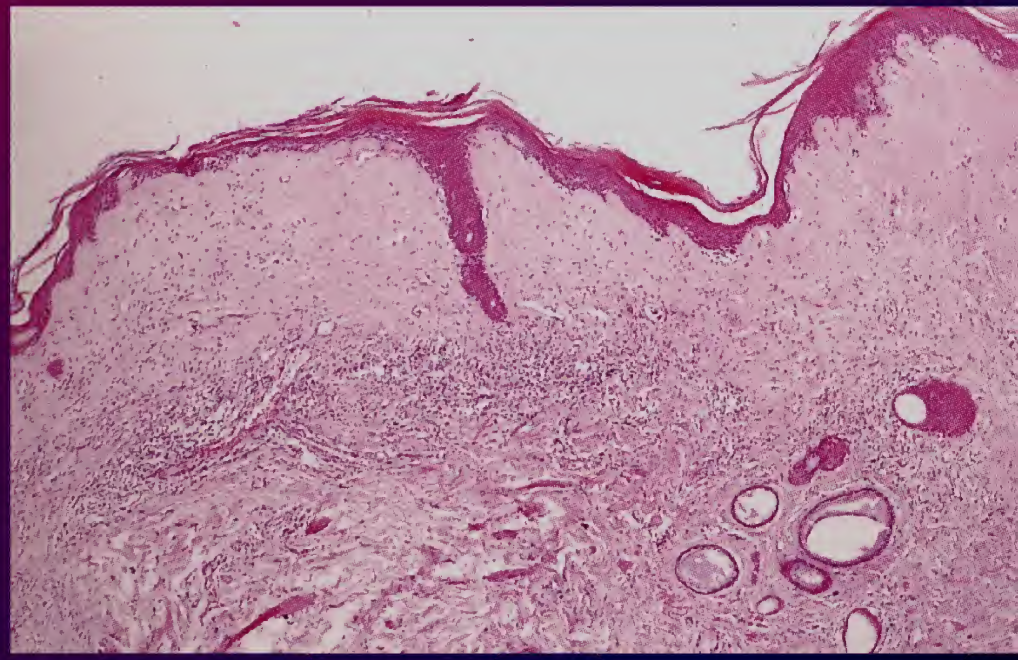
Lichen sclerosus (LS)

- One fourth of women seen in vulva clinics
- 1/300-1/1000 women affected
- Most cases occur postmenopausally
- Can occur in young women
- Equivalent to balanitis xerotica obliterans
- Remittent course
- Can affect non genital skin in 20%
- Autoimmune ? Hormonal ?

Lichen sclerosus

Lichen sclerosus

Lichen sclerosus

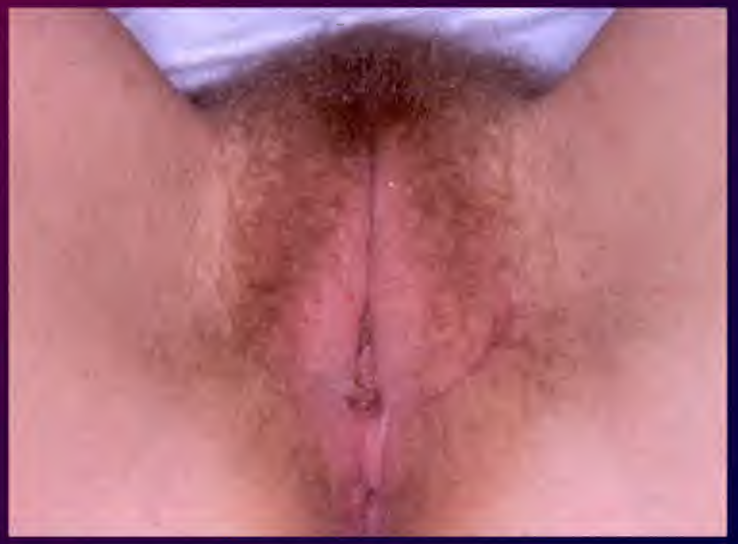


Lichen sclerosus (LS)

- Pruritus is the main symptom
- (3%) associated with vulval carcinoma
- Management is essentially with clobetasol
- Macrolide immunosuppressants (e.g Tacrolimus)
- Surgery is rarely required
- Long term follow up with specialist referral

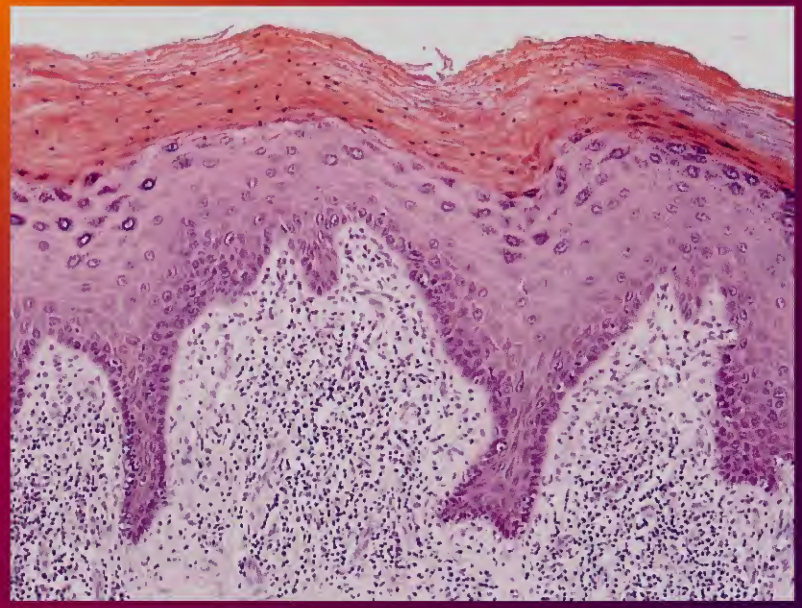
Squamous cell hyperplasia

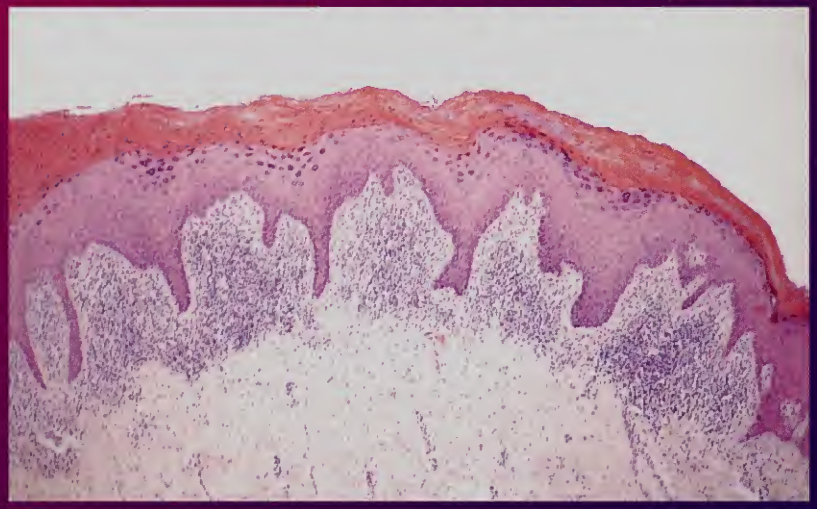
- (Lichen simplex planus)
  (Hyperplastic dystrophy)
- Pruritus plus wide range of skin change.
- Biopsy is essential esp. hyperkeratotic areas
- Long term follow up is needed
- Treatment with steroids



Squamous cell hyperplasia

Squamous cell hyperplasia





Squamous cell hyperplasia

Squamous cell hyperplasia

Squamous cell hyperplasia

- Breaking the itch-scratch cycle is fundamental to the treatment of lichen simplex chronicus (Antihistaminics, Tricyclic antidepressants, SSRIs).
- Steroids
- Treatment of chronic irritative infections

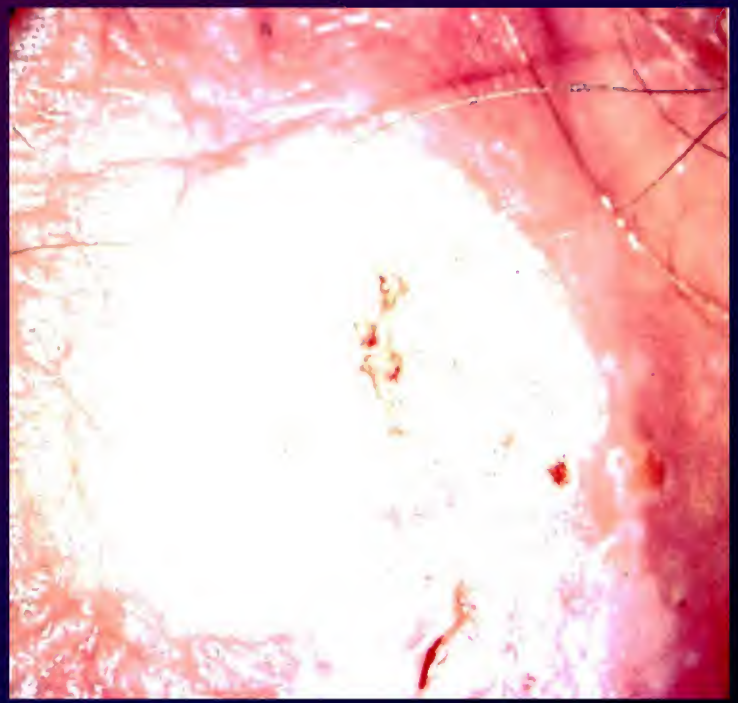
VIN ISSVD classification 2004

- VIN 1 is no longer included
- VIN2 & VIN3 amalgamated in one category

1. **VIN, usual type (HPV related)**
   - VIN, warty
   - VIN, basaloid
   - VIN, mixed
2. **VIN, differentiated type (Non- HPV related)**

VIN 1 now = subclinical HPV

VIN



Vulval intraepithelial neoplasia (VIN)

- Atypia involving the sq. epith.
- Grades according to thickness involved
- Presence of koilocytosis is not an element
- 60% involve non hairy skin
- 66% multifocal and 33% unifocal
- Pruritus, soreness, lump or asymptomatic
- Biopsy & histology are essential

VIN & vulval Paget's

Vulval intraepithelial neoplasia (VIN)

- Younger women usually have multifocal disease
- 43-79% of VIN lesions show HPV
- Risk of invasive disease is obscure (5%)?
- 25-33% of invasive disease show VIN3
- Risk of invasion is usually postmenopausal

Vulval intraepithelial neoplasia (VIN)

Vulval intraepithelial neoplasia (VIN)

- Management involves long term follow up
- Up to 84% recurrence after surgery even in grafted skin
- Biopsy , exclude invasion, expectancy and avoid mutilating surgery
- Treat postmenopausal & immunosuppressed
- Vulvectomy, skinning vulvectomy, WLE, Laser, DNCB, 5-FU, INF, photodynamic therapy
- Immiquimod *van Set ers et al.N Engl J Med 2008;358:1465-73.*

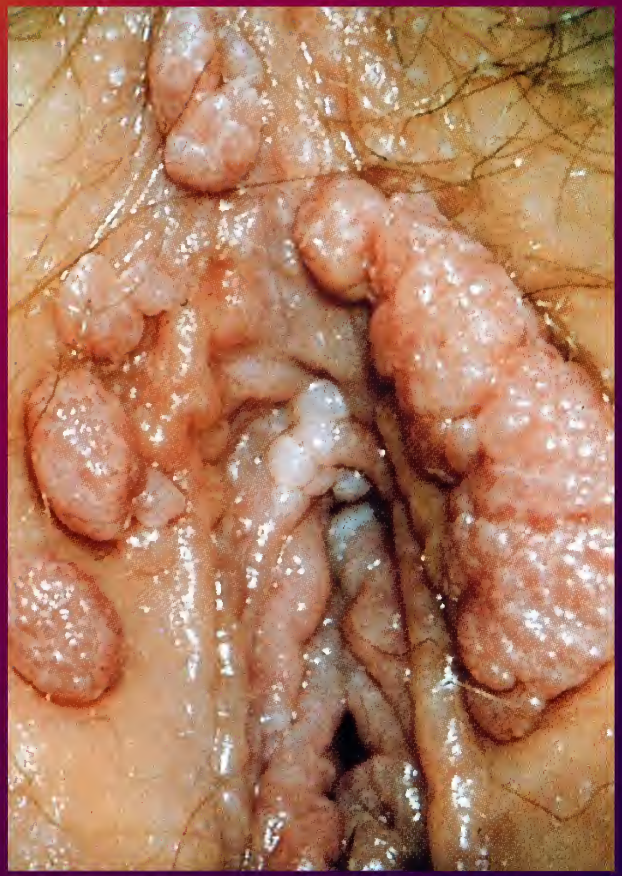
Vulval Paget's

- Sharply demarcated brick red, scaly, eczematoid plaque
- 26% non vulval adenocarcinoma and 4% vulval adenocarcinoma.
- Workup should include colonoscopy, cystoscopy, mammogram, and colposcopy.

Vulval Paget's

- Therapy involves excision with a 2–3 cm safety margin.
- 31% to 43% local recurrence rate
- Laser ablation is not appropriate (need to achieve deep tissue destruction).
- Prognosis is determined by the nature of the coexisting adenocarcinoma, if present.

HPV Condylomata Acuminata



HPV Condylomata Acuminata

- External genital warts caused by HPV 6, 11, 42, 43, 44
- Exophytic benign lesions
- Can cause hyperkeratotic diffuse skin thickening
- Treatment is only for cosmetic purpose

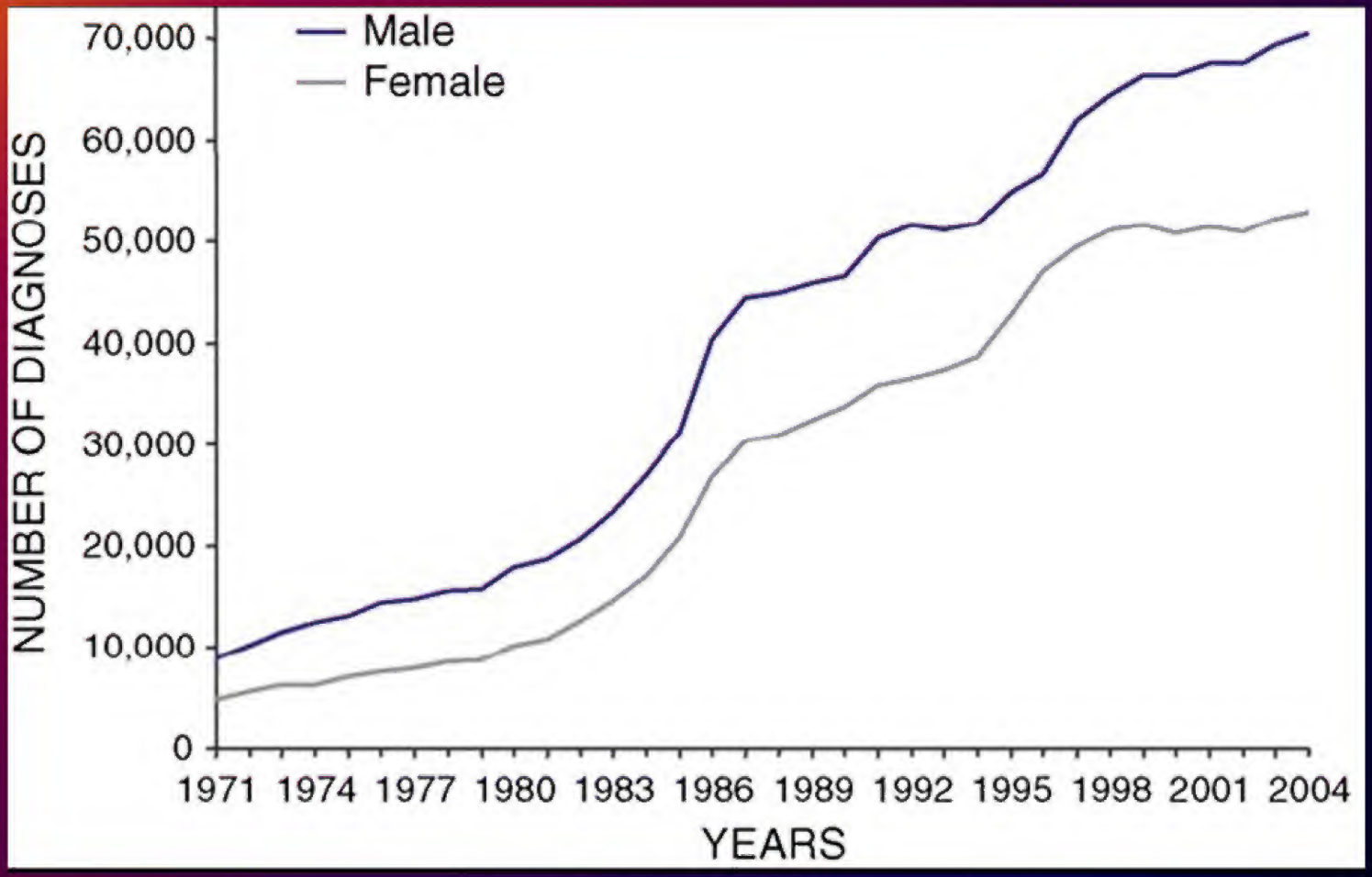
HPV and condyloma accuminata [Genital Warts (GW)]

The UK National Survey of Sexual Attitudes and Lifestyles conducted in the year 2000

- 3.6% of men had Genital warts
- 4.1% of women
- The most common reported sexually transmitted infection.
- Increase over years.

Fenton et al. Lancet 2001;358(9296):1851–4.

Number of diagnoses of genital warts (first, recurrent and reregistered episodes) by sex, STD clinics: England and Wales 1971–2004.



HPV genital warts burden

The cost of a single successful episode of treatment of a case of GWs to be

- $ 377 in the UK ($ 54 million per annum)
- $ 436 in the USA. ($ 200 million per annum)

Langely et al., Int J STD AIDS 2004;15(8):501–8.

Insinga et al., Pharmacoeconomics 2005;23(11):1107–22.

Beware of vestibular papillomatosis

HPV Condylomata Acuminata

- **Cervical Cytology and colposcopy are required**
- **Screen for other STD**
- **Husband or partner screening**

HPV Condylomata Acuminata
Treatment

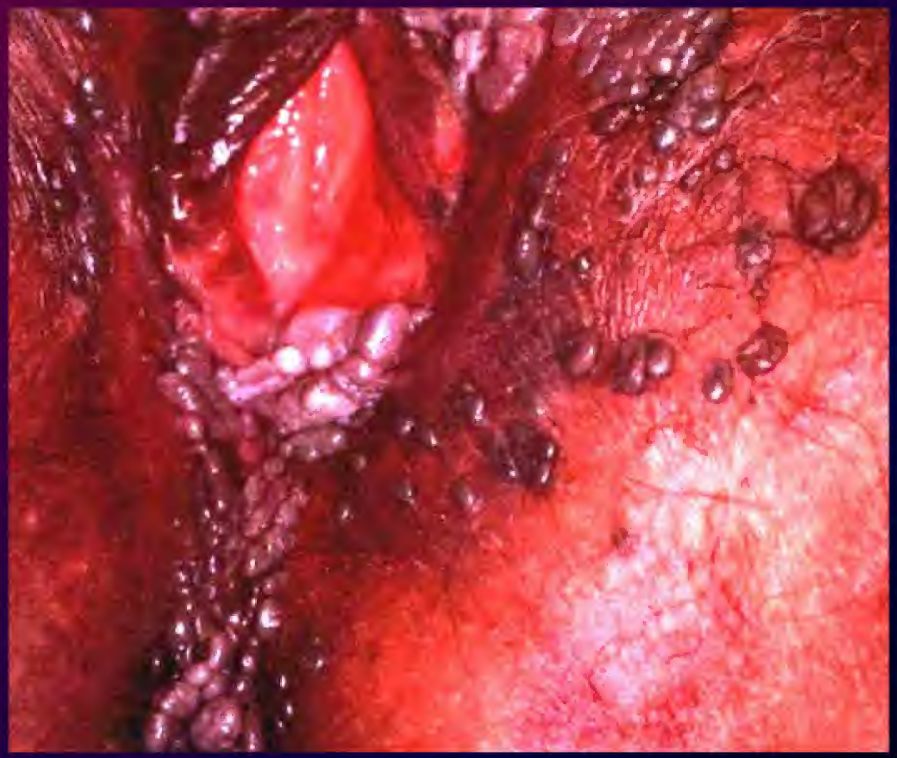
• Podophyllin	22-80%
• BCA and TCA	64-80%
• 5FU	50-90%
• **Cryocautery**	**70-96%**
• **Diathermy**	**72-94%**
• **CO2 Laser**	**72-97%**
• Surgical excision	89-93%
• Systemic interferon	25-35%
• Intralesional interferon	36-52%
• Imiquimod	50%

HPV Condylomata Acuminata

Treatment

Imiquimod 5%

- Aldara cream
- Immune response modifier
- 3 applications weekly for 12-16 week
- 50% success rate



HPV Condyloma Acuminata

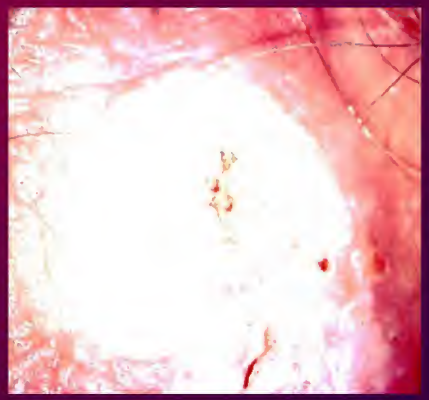
Pregnancy

- Can be treated between 14-32 wks
- Cytotoxic medications can not be used
- Caesarean section doses not alter the risk of vertical transmission
- Caesarean section only when the vagina is involved with multiple vascular growths

Vulval Cancer

- 4-5% of genital malignancy
- 90% squamous cell carcinoma
  - HPV or VIN associated (25-33%)
  - Chronic inflammatory process related
- 2% basal cell carcinoma
- 2-8% melanomas

Vulval cancer



Squamous cell carcinoma of the vulva

FIGO Staging

Stage I: Tumor confined to the vulva

- IA Lesions ≤2 cm in size, confined to the vulva or perineum and with stromal invasion ≤1.0 mm, no nodal metastasis
- IB Lesions >2 cm in size or with stromal invasion >1.0 mm, confined to the vulva or perineum, with negative nodes

Stage II: Tumor of any size with extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with negative nodes

FIGO Staging

Stage III: Tumor of any size with or without extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with positive inguino-femoral lymph nodes

- IIIA (i) With 1 lymph node metastasis (≥5 mm), or
  (ii) 1–2 lymph node metastasis(es) (5 mm)
- IIIB (i) With 2 or more lymph node metastases (≥5 mm), or
  (ii) 3 or more lymph node metastases (<5 mm)
- IIIC With positive nodes with extracapsular spread

FIGO Staging

Stage IV: Tumor invades other regional (2/3 upper urethra, 2/3 upper vagina), or distant structures

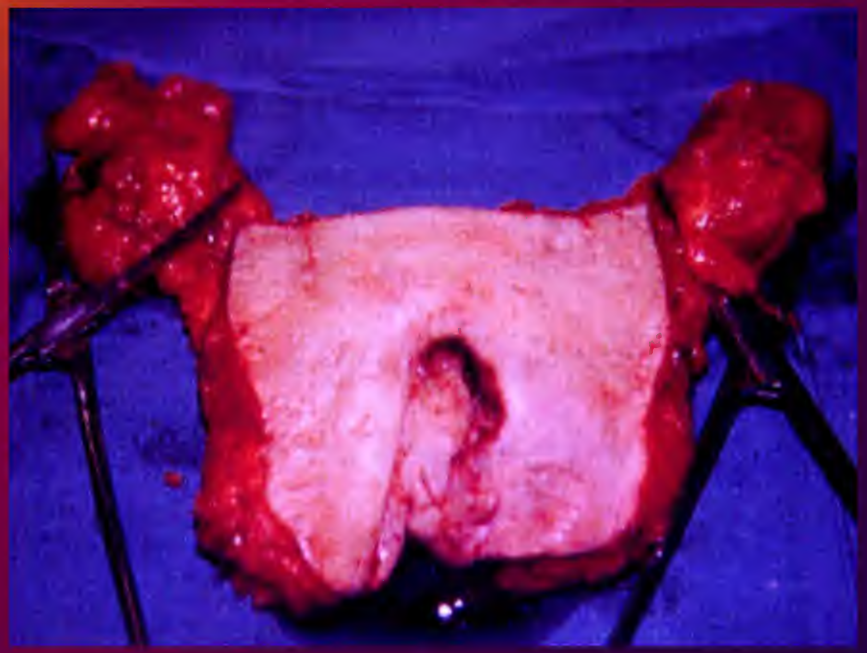
- IVA: Tumor invades any of the following:
  - (i) upper urethral and/or vaginal mucosa, bladder mucosa, rectal mucosa, or fixed to pelvic bone, or
  - (ii) fixed or ulcerated inguino-femoral lymph nodes
- IVB: Any distant metastasis including pelvic lymph nodes

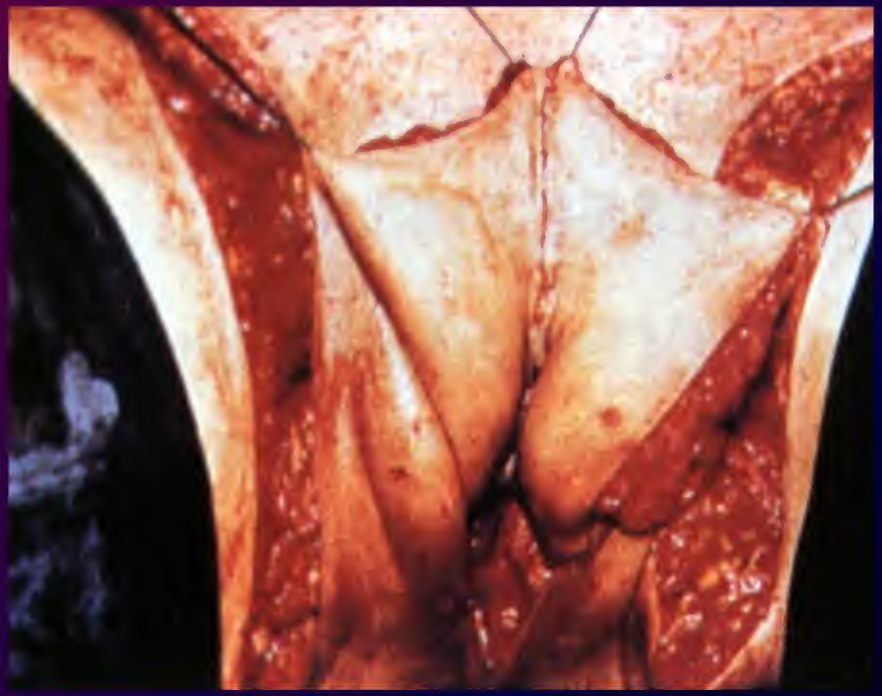
Squamous cell carcinoma of the vulva

Vulval cancer

- Basset's Radical vulvectomy (wound breakdown, TE, lymphoedema)
- Wide local excision and hemivulvectomy
- Split incisions for inguinal node dissection
- Sentinel lymph node technique
- Multimodal therapy and reconstruction for recurrent or advanced disease

Vulval cancer





Conservatism in Vulvar Cancer

This is what we do now

Conservatism in Vulvar Cancer

(*Radical vulvectomy is associated with wound break down, lymphoedema, DVT & TE, mutilation*)

- **Hemivulvectomy + unilateral LND**
- **Split incisions** *(Vs Basset's Butterfly incision)*
- **Sentinel lymph node**
- **Endoscopic inguinal LND**
- **Reconstructive surgery**

Conclusion

- Vulval complaints are not uncommon.
- Vulval complaints can markedly affect patient's quality of life
- Vulval disorders deserve better attention from gynecologists

Conclusion

- Involvement of dermatologists and genito urinary physicians are important.
- Subspecialist opinion (gynecological oncologist) is essential in the management of premalignant and malignant vulval lesions

Acknowledgement

Faculty members, doctors, nursing staff, technicians and workers of

Early Cancer Detection Unit

&

Gynaecological Oncology Unit

Ain Shams University

Thank you.